COARCTATION OF THE AORTA IN AN ADULT, WITH DEATH DUE TO THE RUPTURE OF AN ANEURYSM IN THE NECK

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COARCTATION of the aorta—that is to say, stenosis of the aorta at or just below the junction of the ductus arteriosus with the aorta (i.e., in the region of the aortic "isthmus")—when it is present in adults and in children who have survived the period of infancy, is supposed to have arisen gradually after birth in association with the closure of the ductus arteriosus. Usually in such cases an elaborate system of arterial collateral circulation has developed so that the arterial blood from the upper part of the body—that is to say, from the great arteries given off by the aorta above the site of obstruction—can pass by enlarged anastomosing branches to the lower part of the trunk and the lower extremities. "The blood from above," says A. M. Gossage,[1] "passes by the internal mammary, transverse cervical, superior thoracic and subscapular branches of the subclavians, the long thoracic and subscapular branches of the axillaries, to form anastomosis with the superficial epigastric branch of the femoral and with the thoracic arteries. The blood for the lower limbs is probably mainly conveyed by the thoracic arteries to the abdominal aorta, but owing to the number of small channels pulsation is lost

[1] A. M. Gossage: Proceedings of the Royal Society of Medicine (Clinical Section), 1909, vol. ii., pp. 210-213.

before the abdominal aorta is reached." No wonder, therefore, that in typical cases of the "adult type" a number of thickened tortuous arteries can be both seen and felt pulsating beneath the skin of the back and front of the trunk. The arterial anastomosis may be so efficient that patients with extreme or complete coarctation of the aorta may live to middle age, as in the present case, or even to great old age, and female patients, as in the present case, may be able to rear large families. An early and most interesting observation was that of Reynaud[2] (1828) on a man who, in spite of the aortic coarctation, lived to the advanced age of 92 years.

E. Barié, in 1886, collected together a number of different observations (including Reynaud's case) under the heading, "Du Rétrécissement Congénital de l'Aorte Descendante."[3] In 1903 L. M. Bonnet wrote an elaborate paper on the subject[4] under the heading, "Sur la Lésion dite Sténose Congénitale de l'Aorte dans la Région de l'Isthme." He separated the cases observed in new-born children—that is to say, the "infantile type"—from the other cases—that is to say, the "adult type." Of 160 cases, 55 were of the infantile type and 105 of the adult type. Amongst the adult series there seem to have been 10 cases in which aortic aneurysm of some kind was said to have occurred.

Maud E. Abbott[5] (1908) succeeded in collecting 38 more cases, and by adding these to Bonnet's 160 cases, she obtained a total of 198 cases for her analysis. Of these cases 129 occurred in patients over one year old. Seven patients died in the sixth, and 6 in the seventh decade of life, whilst 1 (Reynaud's case, already alluded to) lived to the age of 92 years. More than half of the remainder (56 cases) died between the ages of 20 and 40 years. In 121 of the 129 cases the sex was known; it was male in 81, female in 40. There was some evidence of collateral circulation in 56 of the 129 cases, but the particular branches involved and the size of the anastomotic arteries

[2] Reynaud: Journal Hebdomadaire de Médecine, Paris, 1828, vol. i., p. 161, illustrated with a plate. The old man's heart was said to be of normal volume.

[3] Barié: Revue de Médecine, Paris, 1886, vol. vi., pp. 343, 408, 501.

[4] Bonnet: Revue de Médecine, Paris, 1903, vol. xxiii., pp. 108, 255, 335, 418, 481.

[5] M. E. Abbott: On Coarctation of the Aorta, in her article on Congenital Cardiac Disease; in Osler and McCrae's System of Medicine, London, 1908, vol. iv., pp. 405–416.

varied greatly even in cases in which the aortic stenosis was extreme. The heart was stated to have remained normal throughout life in 9 of the 129 cases, including Reynaud's case of a man aged 92 years (already referred to); Brunner's case, in which there was an actual obliteration of the aortic channel at the insertion of the ductus arteriosus; and Dumontpallier's case (the patient was aged 39 years), in which the aortic stenosis was due to a septum with a central aperture, 13 mm. in diameter. Seventy-seven cases showed marked hypertrophy and dilatation of the heart, but 44 of them were complicated with chronic valvular lesions or other cardiac defects, which might have given rise to cardiac hypertrophy. Hence it appears that hypertrophy of the heart is not an invariable consequence of aortic coarctation. According to Abbott, aneurysm of the aortic arch occurred in only 9 of the 129 cases, and in 5 of them it was of the dissecting form; rupture of the aorta occurred in 12 cases, of the ascending aorta in 7 cases, and at the site of the stenosis in 5 cases.

Amongst still more recent contributions to the literature of the subject we may mention accounts by A. M. Gossage [6] (a boy, aged 17 years, with characteristic arterial collateral circulation), R. O. Moon [7] (with necropsy), A. Brown [8] (with large tortuous superficial arteries variously disposed over the right thoracic wall), Herbert French [9] (a doubtful case), and Hugo Sella [10] (extreme stenosis of the aortic isthmus in a man, aged 43 years, with perforation of a dissecting aortic aneurysm into the pericardium). George Carpenter [11] has described a case of coarctation of the aorta with great enlargement of superficial arteries in a boy, aged 5 years, who was believed to have inherited syphilis. We should further like to include as an example of the "adult type," with extensive arterial collateral circulation, the case of a woman, aged 55 years, shown by F. Langmead at the Clinical Section of the Royal Society of Medicine on May 31st, 1912, for we cannot think of any other explana-

[6] Gossage: Loc. cit.

[7] R. O. Moon: THE LANCET, June 8th, 1912, p. 1531.

[8] A. Brown: THE LANCET, June 22nd, 1912, p. 1719.

[9] H. French: Transactions of the Medical Society of London, 1911, vol. xxxiv., p. 426.

[10] H. Sella: Ziegler's Beiträge zur Pathologischen Anatomie und zur Allgemeinen Pathologie, Jena, 1910, vol. xlix., p. 501.

[11] G. Carpenter: Reports of the Society for the Study of Disease in Children, London, 1908, vol. viii., p. 158.

tion to account for the presence of the large pulsating superficial arteries on the back of her thorax.[12]

We regard the present case as worthy of record in spite of the fact that no post-mortem examination was obtained, and we have to thank Dr. T. N. Kelynack and Dr. James Mackenzie for their kind permission to publish it. The patient, a Jewish married woman, aged 56 years, was admitted to the Mount Vernon Hospital, Hampstead, on Aug. 24th, 1910. She had had 11 children (seven of them, she said, were still living), the last one 12 years previously, when she herself was 44 years old. She had lived a very active life (housework, &c.), and had apparently enjoyed good health, but since the birth of her last child she had felt less strong. During the last three or four years she had felt weaker, had been getting thinner, and had experienced pain in her chest on exertion. Two years ago she had noticed a lump (doubtless the aneurysm) on the right side of her neck, in front, just above the clavicle. She said that she suffered from occasional feelings of giddiness and headache. She had taken very little alcohol, had never vomited or spat blood, and had not suffered from digestive troubles.

When in the hospital the most remarkable point about the patient was the presence of numerous enlarged and tortuous superficial arteries at various parts of the trunk, neck, and limbs, but especially remarkable in both axillæ and on both sides of the back in the infra-scapular regions. The superficial arteries on the trunk could be felt pulsating under the skin, and some of them were so enlarged as to be nearly as thick as a man's little finger. There was evidently considerable atheromatous change in addition to dilatation and hypertrophy. Above the right clavicle there was a pulsating aneurysmal swelling of about the average size of a hen's egg. Examination of the chest showed the cardiac apex-beat was displaced outwards to the left mid-axillary line in the seventh intercostal space. A systolic murmur could be heard at the apex and over the central and pulmonary regions. The systolic brachial blood pressure varied from 230 to 290 mm. Hg. Dr. Price found that there was an auricular wave in the venous tracing. Roentgen ray examination of the chest showed the aortic arch or part of the aortic arch apparently not continued into

[12] F. Langmead: Proceedings of the Royal Society of Medicine (Clinical Section), London, 1912, vol. v., pp. 194-197 (with discussion).

the descending aorta. The upper part of the heart seemed to be broader than usual. The lungs and diaphragm appeared normal on the screen. No pulsation could be felt in the abdomen to suggest the presence of an abdominal aorta. The abdomen was pendulous owing to flaccidity of its walls. There were varicose veins of the lower extremities (which she thinks developed after the last pregnancy) and dilated veins in front of the chest. Blood examination: hæmoglobin, 92 per cent.; red cells, 5,104,000 in the c.mm. of blood; colour index, 0·8; white cells, 7840 in the c.mm. of blood. The differential count of the white cells gave: polymorphonuclear neutrophiles, 50 per cent.; lymphocytes, 40 per cent.; transitionals, 5 per cent.; eosinophiles, 4 per cent.; and mast cells, 1 per cent. The red cells appeared normal; no nuclear red cells were seen.

The patient was kept a few weeks in the hospital for observation and then went home. After she left the hospital the aneurysmal swelling on the right side of the neck is said to have enlarged until it was nearly as large as a baby's head, and the patient suffered great pain in her right arm (doubtless a pressure effect of the aneurysm). We were informed that she died at her home during the third week of December, 1910. The aneurysm on the side of her neck, after someone had been feeling it, burst externally, the blood squirting out to the other side of the room. She immediately became unconscious and remained so till her death, six hours later. There was no necropsy.